

[illegible]

```

RA Trumbly R.J.: "Cloning and characterization of the CYC8 gene mediating glucose
RT repression in yeast.";
RL Gene 73:97-111(1988).
RN [2]
RP SEQUENCE FROM N.A.
RX MEDLINE=8805502; PubMed=3316983;
RA Schultz J., Carlson M.;
RT "Molecular analysis of SSN6, a gene functionally related to the SNF1
RL protein kinase of Saccharomyces cerevisiae.";
RN Mol. Cell. Biol. 7:3637-3645(1987).
RN [3]
RP SEQUENCE FROM N.A.
RC STRAIN=5288C;
RX MEDLINE=92327848; PubMed=1626431;
RA Mannhaupt G., Stucke R., Ehnlé S., Vetter I., Feldmann H.;
RT "Molecular analysis of yeast chromosome II between CMD1 and LYS2: the
RL excision repair gene RAD16 located in this region belongs to a novel
RT group of double-finger proteins.";
RL Yeast 8:397-408(1992).
RN [4]
RP TPR REPEATS;
RX MEDLINE=90124639; PubMed=2406412;
RA Sivoroski R.S., Boguski M.S., Goebel M., Hieter P.A.;
RT "A repeating amino acid motif in CDC3 defines a family of proteins
RL and a new relationship among genes required for mitosis and RNA
RT synthesis.";
RL Cell 60:307-317(1990).
CC -I- FUNCTION: IT IS INVOLVED IN REPRESSION BY A1-ALPHA2 AND ALPHA2 AND
CC IN OTHER SYSTEMS AS A GENERAL REPRESSOR OF TRANSCRIPTION. THIS
CC PROTEIN HAS NO OBVIOUS DNA-BINDING DOMAINS. IT MIGHT NOT INTERACT
CC DIRECTLY WITH DNA BUT WITH DNA-BOUND PROTEINS.
CC -I- SUBCELLULAR LOCATION: NUCLEAR.
CC -I- SIMILARITY: CONTAINS 10 TPR DOMAINS.
CC -I- SIMILARITY: TO YEAST GAL1 AND CCR4.
CC -----
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CC -----
DR EMBL; M23440; AAA34545.1; -
DR EMBL; M17826; AAA35103.1; -
DR EMBL; X66247; CAA46973.1; -
DR EMBL; X78993; CAA55615.1; -
DR EMBL; Z35981; CAA85069.1; -
DR PIR; S25365; S25365.
DR SGD; S0000316; CYC8.
DR INTERPRO; IPR001440; -.
DR PFAM; PF00515; TPR; 7.
KW Transcription regulation; Repressor; Repeat; TPR domain;
KW Nuclear protein.
FT DOMAIN 15 30 POLY-GLN.
FT REPEAT 46 79 TPR 1.
FT REPEAT 80 113 TPR 2.
FT REPEAT 114 147 TPR 3.
FT REPEAT 150 183 TPR 4.
FT REPEAT 187 220 TPR 5.
FT REPEAT 224 257 TPR 6.
FT REPEAT 258 291 TPR 7.
FT REPEAT 296 329 TPR 8.
FT REPEAT 330 363 TPR 9.
FT REPEAT 364 398 TPR 10.
FT DOMAIN 493 556 30 X 2 AA TANDEM REPEATS OF Q-A.
FT POLY-DOMAIN 557 587 POLY-GLN.
FT CONFLICT 547 547 K -> Q (IN REF. 3).
FO SEQUENCE 966 AA; 107202 MW; 84B509C3208C5C0 CRC64;

```

Query Match

23.78; Score 84; DB 1; Length 966;

[17] VARIANT CAIS ASN-695 AND HIS-695, AND SEQUENCE OF 629-723 FROM N.A.
 RP MEDLINE=921331007; PubMed=1775137;
 RX R15-Stalpers C., Trilliro M.A., Kuiper G.G., Jenster G., Romalo G.,
 RA Sai T., van Rooij H.C., Kaufman M., Rosenfield R.L., Liao S.;
 RT "Substitution of aspartic acid-686 by histidine or asparagine in the
 RL human androgen receptor leads to a functionally inactive protein with
 RM altered hormone-binding characteristics."; *Mol. Endocrinol.* 5:1562-1569(1991).
 RN [18]
 RP VARIANTS CAIS AND PAIS.
 RX MEDLINE=93338440; PubMed=1307250;
 RA Batch J.A., Williams D.M., Davies H.R., Brown B.D., Evans B.A.J.,
 RT Hughes I.A., Patterson M.N.;
 RL "Androgen receptor gene mutations identified by SSCP in fourteen
 RN subjects with androgen insensitivity syndrome."; *Hum. Mol. Genet.* 1:497-503(1992).
 RP [19]
 RP VARIANT CAIS VAL-787.
 RX MEDLINE=92235226; PubMed=1569153;
 RA Nakao R., Hajj M., Yanase T., Ogo A., Takayanagi R., Katsube T.,
 RA Fukumaki Y., Nawata H.;
 RT "A single amino acid substitution (Met-786-->Val) in the steroid-
 RL binding domain of human androgen receptor leads to complete androgen
 RN insensitivity syndrome."; *J. Clin. Endocrinol. Metab.* 74:1152-1157(1992).
 RP [20]
 RP VARIANT LNCAP ALA-877.
 RX MEDLINE=92222955; PubMed=1562599;
 RA Veldscholte J., Berrevoets C.A., Ris-Stalpers C., Kuiper G.G.,
 RA Jenster G., Trapman J., Brinkman A.O., Mulder E.;
 RT "The androgen receptor in LNCap cells contains a mutation in the
 RL ligand binding domain which affects steroid binding characteristics
 RN and response to antiandrogens."; *J. Steroid Biochem. Mol. Biol.* 41:665-669(1992).
 RP [21]
 RP VARIANT MET-730.
 RX MEDLINE=92335289; PubMed=1631125;
 RA Newmark J.R., Hardy D.O., Tonb D.C., Carter B.S., Epstein J.L.,
 RA Isaacs W.B., Brown T.R., Barrack E.R.;
 RT "Androgen receptor gene mutations in human prostate cancer."; *Proc. Natl. Acad. Sci. U.S.A.* 89:6319-6323(1992).
 RN [22]
 RP VARIANT CAIS VAL-754.
 RX MEDLINE=93372806; PubMed=810398;
 RA Lobbaccaro J.-M., Lombroso S., Ktari R., Dumas R., Sultan C.;
 RT "An exonic point mutation creates a Maellit site in the androgen
 RL receptor gene of a family with complete androgen Insensitivity
 RN syndrome."; *Hum. Mol. Genet.* 2:1041-1043(1993).
 RP [23]
 RP VARIANT CAIS ARG-807.
 RX MEDLINE=94108430; PubMed=8281140;
 RA Adegoke O., Kallio P.J., Palvimäki J.J., Kontula K., Jaenue O.A.;
 RT "A single-base substitution in exon 6 of the androgen receptor gene
 RL causing complete androgen insensitivity: the mutated receptor fails
 RN to transactivate but binds to DNA in vitro."; *Hum. Mol. Genet.* 2:1809-1812(1993).
 RP [24]
 RP VARIANT PAIS VAL-743.
 RX MEDLINE=93315568; PubMed=8325932;
 RA Nakao R., Yanase T., Sakai Y., Hajj M., Nawata H.;
 RT "A single amino acid substitution (Gly743-->Val) in the steroid-
 RL binding domain of the human androgen receptor leads to Reifenstein
 RN syndrome."

Query Match	22.3%	Score 79	DB 1	Length 919
Best Local Similarity	32.8%	Pred 0.51		
Matches 19, Conservative 15, Mismatches 22				Indels 2: Gaps 1

QY 5 QQRREEDCRRRCEDQEPHQHQCCLRCREDDQHQHGRGDMNPNQRRGSGRGREYEEGEEDQ 62
 ||::: :::: ||: | : ||::| | : ||: |
 Db 58 QQQQQQ--QQQQQQQQQQQQQQQETSPRQQQQQQQGEDGSGQAHRRCPPTGYLVLDEEDQ 113

ID	NT4_NEUCR	STANDARD:	PRT:	1090 AA.
AC	P28349;			
DT	01-DEC-1992 (Rel. 24, Created)			
DT	01-DEC-1992 (Rel. 24, Last sequence update)			
DT	01-NOV-1997 (Rel. 35, Last annotation update)			
DE	NITROGEN ASSIMILATION TRANSCRIPTION FACTOR NIT-4.			
GN	NIT-4.			
OS	Neurospora crassa.			
OC	Eukaryota; Fungi; Ascomycota; Sordariales; Sordariaceae; Neurospora.			
RN	[1]			
RP	SEQUENCE FROM N.A.			
RX	MEDLINE=92017855; PubMed=1840634;			
RA	Yuan G.-F., Fu Y.-H., Marzluff G.A.;			
RT	"nit-4, a pathway-specific regulatory gene of Neurospora crassa,			
RT	encodes a protein with a putative binuclear zinc DNA-binding			
RL	domain.";			
RL	Mol. Cell. Biol. 11:5735-5745(1991).			
RN	[2]			
RP	SEQUENCE FROM N.A.			
RX	MEDLINE=92149315; PubMed=1531376;			
RA	Yuan G.-F., Marzluff G.A.;			
RT	"molecular characterization of mutations of nit-4, the			
RT	pathway-specific regulatory gene which controls nitrate assimilation			
RL	in Neurospora crassa.";			
RL	Mol. Microbiol. 6:67-73(1992).			
CC	-1- FUNCTION: PATHWAY-SPECIFIC REGULATORY GENE OF NITRATE			
CC	ASSIMILATION: IT ACTIVATES THE TRANSCRIPTION OF THE GENES FOR			
CC	NITRATE AND NITRITE REDUCTASES.			
CC	-1- SUBCELLULAR LOCATION: NUCLEAR.			
CC	-1- DOMAIN: THE GLUTAMINE-RICH DOMAIN MIGHT FUNCTION IN ACTIVATING			
CC	GENE EXPRESSION.			
CC	-1- SIMILARITY: CONTAINS A ZN(2)-CYS(6), FUNGAL-TYPE BINUCLEAR			
CC	CLUSTER DOMAIN.			
CC	-----			
CC	This SWISS-PROT entry is copyright. It is produced through a collaboration			
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CC	or send an email to license@isb-sib.ch).			
CC	-----			
DR	EMBL: M80368; AAA33602.1; -			
DR	PIR: A41696; A41696.			
DR	PIR: S20033; S20033.			
DR	HSSP: P07272; IPTI.			
DR	INTERPRO: IPR001138; -			
DR	PFAM: PF001172; Zn_C1us; 1.			
DR	PROSITE: PS00463; ZN2_Cy6; FUNGAL_1; 1.			
DR	PROSITE: PS50048; ZN2_Cy6; FUNGAL_2; 1.			
KW	Transcription regulation; Activator; DNA-binding; Nuclear protein;			
KW	Zinc; Metal-binding; Nitrate assimilation.			
FT	DNA_BIND	53	81	
FT	DOMAIN	121	139	
FT	DOMAIN	213	229	
FT	DOMAIN	429	450	
FT	DOMAIN	672	754	
FT	DOMAIN	755	859	
FT	DOMAIN	992	1024	
FT	CONFLICT	98	98	
FT	CONFLICT	467	467	
QO	SEQUENCE	1090 AA;	120244 MW;	861DB9172EDD6114 CRC64;

```

Query Match      21.1%; Score 75; DB 1; Length 1090;
Best Local Similarity 32.7%; Pred. No. 1.5;
Matches 17; Conservative 14; Mismatches 17; Indels 4; Gaps 1;

Oy      6 GREEDCRRRCDEQEPROHQCQLRCREGRORHCRGDDMM----NPGRSGS 53
       11::|::||::||| | | ::|||:| : :|

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OS Mus musculus (Mouse).
OC Eukaryota; Metazoa; Chordata; Vertebrata; Euteleostomi;
OC Mammalia; Eutheria; Rodentia; Sciurognathi; Muridae; Murinae; Mus.
RN (1)
RN SEQUENCE FROM N.A.
RC TISSUE-LYMPHOMA;
RA Fisher R., Fillmore H., Reynolds A.B.;
RT "Molecular cloning and characterization of the 162 kDa component
of a multi-protein complex phosphorylated by Src.";
RL Submitted (SEP-1994) to the EMBL/GenBank/DBJ databases.
RN (2)
RP SEQUENCE OF 514-961 FROM N.A. (CENTROSOMIN B).
RA MEDLINE=98039715; PubMed=9372446;
RX Petzelt C., Joswig G., Mincheva A., Licher P., Stammer H., Werner D.;
RT "The centrosomal protein centrosomin A and the nuclear protein
centrosomin B derive from one gene by post-transcriptional processes
involving RNA editing.";
RL J. Cell Sci. 110:2573-2578(1997).
RN (3)
RP SEQUENCE OF 514-790 FROM N.A. (CENTROSOMIN A).
RA MEDLINE=91277032; PubMed=1829085;
RX Joswig G., Petzelt C., Werner D.;
RT "Murine cDNAs coding for the centrosomal antigen centrosomin A.";
RL J. Cell Sci. 98:37-43(1991).
RN (4)
RN REVISIONS.
RA Joswig G., Petzelt C., Werner D.;
RL Submitted (DEC-1996) to the EMBL/GenBank/DBJ databases.
CC -1- FUNCTION: BINDS TO THE 40S RIBOSOME AND PROMOTES THE BINDING OF
METHIONYL-TRNAI AND MRNA.
CC -1- SUBUNIT: EFF-3 IS COMPOSED OF AT LEAST 10 DIFFERENT SUBUNITS.
CC -1- SUBCELLULAR LOCATION: CYTOPLASMIC (BY SIMILARITY).
CC -1- PTM: PHOSPHORYLATED.
CC -1- SIMILARITY: BELONGS TO THE EIF3S10 FAMILY.
CC -----
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CC -----
DR EMBL: U14172; AAA09010.1; -;
DR EMBL: X84651; CAA59144.1; -;
DR EMBL: X17373; CAA35246.1; -;
DR PIR: S13800; S13800.
DR MGD: MGI:95301; EIF3.
KW Initiation factor; Protein biosynthesis; Repeat; Phosphorylation.
FT DOMAIN 924 1133
FT 21 X 10 AA TANDM REPEAT OF D-[DE]-D-R-
FT CONFLICT 613 647
FT [GP]-[PS]-[RW]-R-[GN]-[AM].
FT ROEAKEREKRILOEHEQIKKTVRELRQIKKTE ->
FT PRKGAREGENTNSRTFRANGECGAVRADQEDR (IN
FT REF. 2 AND 3).
FT CONFLICT 683 684
FT EL -> DY (IN REF. 2 AND 3).
FT CONFLICT 717 717
FT Q -> H (IN REF. 2 AND 3).
FT CONFLICT 766 766
FT A -> V (IN REF. 2 AND 3).
FT CONFLICT 787 790
FT RHR -> SIVA (IN REF. 3).
FT CONFLICT 793 793
FT E -> D (IN REF. 2).
SO SEQUENCE 1344 AA; 161949 MW; F4CAE2169F577712 CRC64;

Query Match 19.9%; Score 70.5; DB 1; Length 1344;
Best Local Similarity 26.4%; Pred. No. 5.2;
Matches 19; Conservative 20; Mismatches 24; Indels 9; Gaps 3;

OY 1 KRPDPRREYEDCRRRCEQDF-PRQGHQCLRCRCEQQRQGR--GGMMNPQ-----RGS 51
DB 836 KKEELRREYQERVKKLEVERKKRRELEIERRRERREERRLGDDPLSRKDSRWGDRDS 895
OY 52 SGRVEGEEES 63
DB 896 EGTWRKGPEDAS 907

RESULT 12
WT1_SMIMA STANDARD; PRT; 239 AA.
ID WT1_SMIMA
AC P49953;
DT 01-OCT-1996 (Rel. 34, Created)
DT 01-OCT-1996 (Rel. 34, Last sequence update)
DT 01-OCT-2000 (Rel. 40, Last annotation update)
DE WT1MS' TUMOR PROTEIN (FRAGMENT).
GN WT1.
OS Sinthopsis macronura (Stripe-faced dunart).
OC Eukaryota; Metazoa; Chordata; Craniata; Vertebrata; Euteleostomi;
OC Mammalia; Metatheria; Dasyuromorpha; Dasyuridae; Sinthopsis.
RN (1)
RN SEQUENCE FROM N.A.
RC TISSUE-TESTIS;
RA MEDLINE=96068905; PubMed=7478606;
RX Kent J., Coriat A.M., Sharpe P.T., Hastie N., van Heyningen V.;
RT "The evolution of WT1 sequence and expression pattern in the
vertebrates.";
RL Oncogene 11:1781-1792(1995).
CC -1- FUNCTION: POTENTIAL ROLE IN TRANSCRIPTIONAL REGULATION. RECOGNIZES
AND BINDS TO THE DNA SEQUENCE 5'-CGCCCCCG-3' (BY SIMILARITY).
CC -1- SUBCELLULAR LOCATION: NUCLEAR.
CC -1- ALTERNATIVE PRODUCTS: TWO ALTERNATIVE SPLICE SITES EXISTS (BY
SIMILARITY).
CC -1- SIMILARITY: BELONGS TO THE EGR FAMILY OF C2H2-TYPE ZINC-FINGER
PROTEINS.
CC -----
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CC -----
DR EMBL: X85732; CAA59737.1; -;
DR HSSP: P08046; IAL1.
DR INTERPRO: IPR000822; -;
DR PIR: PR00096; zfc2H2; 4.
DR PROSITE: PS00028; ZINC_FINGER_C2H2; 4.
KW Zinc-finger; Metal-binding; DNA-binding; Repeat; Nuclear protein;
KW Transcription regulation; Alternative splicing; Anti-oncogene.
FT NON_TER 1 1
FT DOMAIN 113 228
FT ZN_FING 113 137
FT ZN_FING 143 167
FT ZN_FING 173 195
FT ZN_FING 204 228
FT VARSPIC 198 200
FT MISSING (IN ISOFORM 2).
SO SEQUENCE 239 AA; 27793 MW; 6707678A7259A624 CRC64;

Query Match 19.4%; Score 69; DB 1; Length 239;
Best Local Similarity 25.0%; Pred. No. 1.5;
Matches 15; Conservative 14; Mismatches 25; Indels 6; Gaps 1;

OY 4 PQOREYEDCRRRCEQDEPRQHQCO-----LRCRQDQHQHGGMMNPQSGSRTEE 57
DB 142 PYQCDKDCRRRPSRSDQLKRHQRRHTGVKPPQCKTKRFSKSHLKTHTTHTGKTSJ 201

RESULT 13
WT1_ALIMI STANDARD; PRT; 288 AA.
ID WT1_ALIMI
AC P50902;
DT 01-OCT-1996 (Rel. 34, Created)
DT 01-OCT-1996 (Rel. 34, Last sequence update)
DT 01-OCT-2000 (Rel. 40, Last annotation update)
DE WT1MS' TUMOR PROTEIN (FRAGMENT).
GN WT1.


```

OS Alligator mississippiensis (American alligator).
OC Eukaryota; Metazoa; Chordata; Cranialata; Vertebrata; Euteleostomi;
OC Actinoptera; Crocodylidae; Alligatorinae; Alligator.
RN [1]
RP SEQUENCE FROM N.A.
RX MEDLINE=96068905; Pubmed=7478606;
RA Kent J., Coriat A.M., Sharpe P.T., Hastie N., van Heyningen V.;
RT "The evolution of Wt1 sequence and expression pattern in the
RT vertebrates."
RL Oncogene 11:1781-1792(1995).
CC -1- FUNCTION: POTENTIAL ROLE IN TRANSCRIPTIONAL REGULATION. RECOGNIZES
CC AND BINDS TO THE DNA SEQUENCE 5'-CCCCCCCC-3'.
CC -1- SUBCELLULAR LOCATION: NUCLEAR.
CC -1- ALTERNATIVE PRODUCTS: TWO ALTERNATIVE SPLICED SITES EXISTS (BY
CC SIMILARITY).
CC -1- SIMILARITY: BELONGS TO THE EGR FAMILY OF C2H2-TYPE ZINC-FINGER
CC PROTEINS.
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-----
DR EMBL; X85730; CAA59735.1; -.
DR HSSP; P08046; 1A1L.
DR INTERPRO; IPR000822; -.
DR PRAM; PF00096; zf-C2H2; 4.
DR PROSITE; PS00028; ZINC_FINGER_C2H2; 4.
KW Zinc-finger; Metal-binding; DNA-binding; Repeat; Nuclear protein;
KW Transcription regulation; Alternative splicing.
FT NCN_TER 1
FT ZN_FING 162 186 C2H2-TYPE.
FT ZN_FING 192 216 C2H2-TYPE.
FT ZN_FING 222 244 C2H2-TYPE.
FT ZN_FING 253 277 C2H2-TYPE.
FT VARSPIC 245 247 MISSING (IN ISOFORM 2).
SO SEQUENCE 288 AA; 33111 MW; 33E26F7DBE7BFDE CRC64;

Query Match 19.4%; Score 69; DB 1; Length 288;
Best Local Similarity 25.0%; Pred. No. 1.8;
Matches 15; Conservative 14; Mismatches 25; Indels 6; Gaps 1;

OY 4 POOREYEDCRRRCRQGPPOOHQO-----LRCREOQROHGRGDMNORGGSGRYEE 57
DB 191 PYOCDFDCRRRSRSDQLKRRHRTGVRPQCKTCQKRFSDHLKTRHTHTGKTS 250

RESULT 14
WT1_RAT ID WT1_RAT STANDARD; PRT; 448 AA.
AC P49552;
DT 01-OCT-1996 (Rel. 34, Created)
DT 01-OCT-1996 (Rel. 34, Last sequence update)
DT 01-OCT-2000 (Rel. 40, Last annotation update)
DE WILMS' TUMOR PROTEIN HOMOLOG.
GN WT1 OR WT-1.
OS Rattus norvegicus (Rat).
OC Eukaryota; Metazoa; Chordata; Cranialata; Vertebrata; Euteleostomi;
OC Mammalia; Eutheria; Rodentia; Sciurognathi; Muridae; Murinae; Rattus.
RN [1]
RP SEQUENCE FROM N.A.
RX STRAIN=SPRAGUE-DAWLEY; TISSUE=KIDNEY;
RX MEDLINE=93046155; Pubmed=1330293;
RA Sharma P.M., Yang X., Bowman M., Roberts V., Sukumar S.;
RT "Molecular cloning of rat Wt1ms' tumor complementary DNA and a study
RT of messenger RNA expression in the urogenital system and the brain."
RL Cancer Res. 52:6407-6412(1992).
CC -1- FUNCTION: POTENTIAL ROLE IN TRANSCRIPTIONAL REGULATION. RECOGNIZES
CC AND BINDS TO THE DNA SEQUENCE 5'-GCCCCCCC-3'.

```

```

CC -1- SUBCELLULAR LOCATION: NUCLEAR.
CC -1- ALTERNATIVE PRODUCTS: TWO ALTERNATIVE SPLICED SITES EXISTS.
CC -1- TISSUE SPECIFICITY: KIDNEY (BY SIMILARITY).
CC -1- DEVELOPMENTAL STAGE: EXPRESSED DURING KIDNEY DEVELOPMENT.
CC -1- SIMILARITY: BELONGS TO THE EGR FAMILY OF C2H2-TYPE ZINC-FINGER
CC PROTEINS.
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-----
DR EMBL; X69716; CAA49373.1; -.
DR TRANSFAC; T02352; -.
DR INTERPRO; IPR000822; -.
DR PRAM; PF00096; zf-C2H2; 4.
DR PRINTS; PR00048; ZINC_FINGER.
DR PROSITE; PS00049; WILMS_TUMOR.
DR PROSITE; PS00028; ZINC_FINGER_C2H2; 4.
KW Zinc-finger; Metal-binding; DNA-binding; Repeat; Nuclear protein;
KW Transcription regulation; Alternative splicing; Anti-oncogene.
FT DOMAIN 27 82 ZINC_FINGERS.
FT ZN_FING 322 437 C2H2-TYPE.
FT ZN_FING 352 376 C2H2-TYPE.
FT ZN_FING 382 404 C2H2-TYPE.
FT ZN_FING 413 437 C2H2-TYPE.
FT VARSPIC 249 265 MISSING (IN ISOFORM 2 AND ISOFORM 3).
FT VARSPIC 407 409 MISSING (IN ISOFORM 2 AND ISOFORM 4).
SO SEQUENCE 448 AA; 49193 MW; 329AC9AC1FF73F76 CRC64;

Query Match 19.4%; Score 69; DB 1; Length 448;
Best Local Similarity 25.0%; Pred. No. 2.7;
Matches 15; Conservative 14; Mismatches 25; Indels 6; Gaps 1;

OY 4 POOREYEDCRRRCRQGPPOOHQO-----LRCREOQROHGRGDMNORGGSGRYEE 57
DB 351 PYOCDFDCRRRSRSDQLKRRHRTGVRPQCKTCQKRFSDHLKTRHTHTGKTS 410

RESULT 15
WT1_HUMAN ID WT1_HUMAN STANDARD; PRT; 449 AA.
AC P19544;
DT 01-FEB-1991 (Rel. 17, Created)
DT 01-AUG-1991 (Rel. 19, Last sequence update)
DT 01-OCT-2000 (Rel. 40, Last annotation update)
DE WILMS' TUMOR PROTEIN (WT33).
GN WT1.
OS Homo sapiens (Human).
OC Eukaryota; Metazoa; Chordata; Cranialata; Vertebrata; Euteleostomi;
OC Mammalia; Eutheria; Primates; Catarrhini; Homnidae; Homo.
RN [1]
RP SEQUENCE FROM N.A.
RX TISSUE=RETAL KIDNEY;
RX MEDLINE=90158822; Pubmed=2154702;
RA Gessler M., Poustka A., Cavenee W., Neve R.L., Orkin S.H.,
RA Bruns G.A.P.;
RT "Homologous deletion in Wt1ms tumours of a zinc-finger gene
RT identified by chromosome jumping."
RL Nature 343:774-778(1990).
RN [2]
RP SEQUENCE FROM N.A.
RX TISSUE=PLACENTA;
RX MEDLINE=92052142; Pubmed=1658787;
RA Haber D.A., Sohn R.L., Buckler A.J., Pelletier J., Call K.M.,
RA Housman D.E.;
RT "Alternative splicing and genomic structure of the Wt1ms tumor gene

```

RT WTI.";
 RL Proc. Natl. Acad. Sci. U.S.A. 88:9618-9622(1991).
 RN [3]
 RX SEQUENCE OF 85-449 FROM N.A.
 RP MEDLINE=90150277; PubMed=2154335;
 RA Call K.M., Glaser T., Ito C.Y., Buckler A.J., Pelletier J.,
 RA Haberman D.E.,
 RA Haberman D.A., Rose E.A., Kral A., Yeger H., Lewis W.H., Jones C.,
 RT "Isolation and characterization of a zinc finger polypeptide gene at
 RT the human chromosome 11 Wilms' tumor locus.";
 RL Cell 60:509-520(1990).
 RN [4]
 RP IDENTIFICATION OF START CODON AND ALTERNATIVE SPLICING SITES.
 RX MEDLINE=91141522; PubMed=1671709;
 RA Buckler A.J., Pelletier J., Haber D.A., Glaser T., Housman D.E.,
 RT "Isolation, characterization, and expression of the murine Wt1"
 RT tumor gene (WT1) during kidney development.";
 RL Mol. Cell. Biol. 11:1707-1712(1991).
 RN [5]
 RP VARIANT WT CYS-366.
 RX MEDLINE=92279213; PubMed=1317572;
 RA Little M.H., Prosser J., Condie A., Smith P.J., van Heyningen V.,
 RT "Zinc finger point mutations within the WT1 gene in Wilms tumor
 RT patients.";
 RL Proc. Natl. Acad. Sci. U.S.A. 89:4791-4795(1992).
 RN [6]
 RP VARIANTS DDS.
 RX MEDLINE=92005721; PubMed=1655284;
 RA Pelletier J., Bruening W., Kashan C.E., Mauer S.M., Manivel J.C.,
 RA Striegel J.E., Houghton D.C., Junien C., Habib R., Fouser L.,
 RA Fine R.N., Silverman B.L., Haber D.A., Housman D.,
 RT "Germline mutations in the Wt1 tumor suppressor gene are
 RT associated with abnormal urogenital development in Denys-Drash
 RT syndrome.";
 RL Cell 67:437-447(1991).
 RN [7]
 RP VARIANTS DDS.
 RX MEDLINE=93265053; PubMed=1338906;
 RA Baird P.N., Santos A., Groves N., Jadresic L., Cowell J.K.,
 RT "Constitutional mutations in the WT1 gene in patients with
 RT Denys-Drash syndrome.";
 RL Hum. Mol. Genet. 1:301-305(1992).
 RN [8]
 RP VARIANTS DDS.
 RX MEDLINE=93271983; PubMed=8388765;
 RA Little M.H., Williamson K.A., Mannens M., Kelsey A., Gosden C.,
 RT "Evidence that WT1 mutations in Denys-Drash syndrome patients may act
 RT in a dominant-negative fashion.";
 RL Hum. Mol. Genet. 2:259-264(1993).
 RN [9]
 RP VARIANT MESOTHELIOMA GLY-273.
 RX MEDLINE=94004972; PubMed=8401592;
 RA Park S., Schalling M., Bernard A., Maheswaran S., Shipley G.C.,
 RA Roberts D., Fletcher J., Shipman R., Rheinwald J., Demetri G.,
 RA Griffin J., Minden M., Housman D.E., Haber D.A.,
 RT "The Wt1 tumor gene WT1 is expressed in murine mesoderm-derived
 RT tissues and mutated in a human mesothelioma.";
 RL Nat. Genet. 4:415-420(1993).
 RN [10]
 RP VARIANTS WT SER-181 AND ALA-253.
 RX MEDLINE=97268681; PubMed=9108089;
 RA Schumacher V., Schneider S., Figge A., Wildhardt G., Harms D.,
 RT Schmidt D., Weirich A., Ludwig R., Royer-Pokora B.,
 RT "Correlation of germ-line mutations and two-hit inactivation of the
 RT WT1 gene with Wilms tumors of stromal-predominant histology.";
 RL Proc. Natl. Acad. Sci. U.S.A. 94:3972-3977(1997).
 RN [11]
 RP VARIANTS DMS TYR-377; LEU-383 AND ASN-396.
 RX MEDLINE=98198341; PubMed=9529364;
 RA Jeanpierre C., Denamur E., Henry I., Cabanis M.-O., Luce S.,
 RA Cecille A., Elion J., Peuchmaur M., Lohrat C., Naudet P.,

RA Gubler M.-C., Junien C.,
 RT "Identification of constitutional WT1 mutations, in patients with
 RT isolated diffuse mesangial sclerosis, and analysis of
 RT genotype/phenotype correlations by use of a computerized mutation
 RT database.";
 RL Am. J. Hum. Genet. 62:824-833(1998).
 RN [12]
 RP REVIEW.
 RX MEDLINE=92207913; PubMed=1313285;
 RA Haber D.A., Buckler A.J.,
 RT "WT1: a novel tumor suppressor gene inactivated in Wilms' tumor.";
 RL New Biol. 4:97-106(1992).
 RN [13]
 RP REVIEW.
 RX MEDLINE=93345769; PubMed=8393820;
 RA Rauscher F.J. III,
 RT "The WT1 Wilms tumor gene product: a developmentally regulated
 RT transcription factor in the kidney that functions as a tumor
 RT suppressor.";
 RL FASEB J. 7:896-903(1993).
 CC -1- FUNCTION: POTENTIAL ROLE IN TRANSCRIPTIONAL REGULATION. RECOGNIZES
 CC AND BINDS TO THE DNA SEQUENCE 5'-CGCCCCCC-3'.
 CC -1- SUBCELLULAR LOCATION: NUCLEAR.
 CC -1- ALTERNATIVE PRODUCTS: TWO ALTERNATIVE SPLICING SITES EXISTS.
 CC -1- TISSUE SPECIFICITY: EXPRESSED IN THE KIDNEY AND A SUBSET OF
 CC HEMATOPOIETIC CELLS.
 CC -1- DISEASE: WILMS TUMOR (WT) IS AN EMBRYONAL MALIGNANCY OF THE
 CC KIDNEY THAT AFFECTS APPROXIMATELY 1 IN 10,000 INFANTS AND YOUNG
 CC CHILDREN. IT OCCURS BOTH IN SPORADIC AND HEREDITARY FORMS.
 CC -1- DISEASE: PATIENTS WITH DENYS-DRASH SYNDROME (DDS), WHICH IS
 CC CHARACTERIZED BY A TYPICAL NEPHROPATHY AND GENITAL ABNORMALITIES,
 CC HAVE DEFECTS IN THE WT1 GENE.
 CC -1- DISEASE: DEFECTS IN WT1 ARE ALSO A CAUSE OF DIFFUSE MESANGIAL
 CC SCLEROSIS (DMS), A FORM A DDS.
 CC -1- SIMILARITY: BELONGS TO THE EGR FAMILY OF C2H2-TYPE ZINC-FINGER
 CC PROTEINS.
 CC -1- DATABASE: NAME=WT1; NOTE=WT1 mutation database;
 CC WWW="http://www.umd.necker.fr:2003/".
 CC -----
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 DR EMBL: X51630; CAA35956.1; ALT_INIT.
 DR EMBL: M80232; AAA61299.1; -
 DR EMBL: M80217; AAA61299.1; JOINED.
 DR EMBL: M80218; AAA61299.1; JOINED.
 DR EMBL: M80219; AAA61299.1; JOINED.
 DR EMBL: M80220; AAA61299.1; JOINED.
 DR EMBL: M80221; AAA61299.1; JOINED.
 DR EMBL: M80228; AAA61299.1; JOINED.
 DR EMBL: M80229; AAA61299.1; JOINED.
 DR EMBL: M80231; AAA61299.1; JOINED.
 DR EMBL: M30393; AAA36810.1; -
 DR PIR: A34673; A34673.
 DR PIR: S08273; S08273.
 DR TRNSFAC: T00899; -
 DR MIM: 194070; -
 DR MIM: 194080; -
 DR MIM: 256370; -
 DR INTERPRO: IPR000822; -
 DR INTERPRO: IPR000976; -
 DR PEFAM: PFO00096; zf-C2H2; 4.
 DR PRINTS: PR00048; ZINCINGER.
 DR PRINTS: PR00049; WILMS_TUMOR.
 DR PROSITE: PS00028; ZINC_FINGER_C2H2; 4.
 DR Zinc-finger: Metal-binding; DNA-binding; Repeat; Nuclear protein;
 KW Transcription regulation; Alternative splicing; Anti-oncogene;

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KW Disease mutation.
FT DOMAIN 27 83 PRO-RICH.
FT DOMAIN 323 438 ZINC FINGERS.
FT ZN_FING 323 347 C2H2-TYPE.
FT ZN_FING 353 377 C2H2-TYPE.
FT ZN_FING 383 405 C2H2-TYPE.
FT ZN_FING 414 438 C2H2-TYPE.
FT VARSPLIC 250 266 MISSING (IN ISOFORM 2 AND ISOFORM 3).
FT VARSPLIC 408 410 MISSING (IN ISOFORM 2 AND ISOFORM 4).
FT VARIANT 181 181 P -> S (IN WT).
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FT VARIANT 223 223 S -> N (IN WT).
FT VARIANT 223 223 /FTId=VAR_007740.
FT VARIANT 253 253 G -> A (IN WT).
FT VARIANT 253 253 /FTId=VAR_007741.
FT VARIANT 273 273 S -> G (IN MESOTHELIOMA).
FT VARIANT 273 273 /FTId=VAR_007742.
FT VARIANT 330 330 C -> Y (IN DDS).
FT VARIANT 330 330 /FTId=VAR_007743.
FT VARIANT 360 360 C -> G (IN DDS).
FT VARIANT 360 360 /FTId=VAR_007744.
FT VARIANT 366 366 R -> C (IN WT).
FT VARIANT 366 366 /FTId=VAR_007745.
FT VARIANT 366 366 R -> H (IN DDS).
FT VARIANT 366 366 /FTId=VAR_007746.
FT VARIANT 373 373 H -> Q (IN DDS).
FT VARIANT 373 373 /FTId=VAR_007747.
FT VARIANT 377 377 H -> Y (IN DDS/DMS).

Query Match 19.4%; Score 69; DB 1; Length 449;
Best Local Similarly 25.0%; Pred. No. 2.7;
Matches 15; Conservative 14; Mismatches 25; Indels 6; Gaps 1; .

QY 4 POREVEDCRRCRCEQGEPROQHOC-----LRCREQQRQGRGSDMNPQRGSSGRYEE 57
| | : : : | | | : : : | | : : : | | : : : | | : : : |
Db 352 PYQDFRDCERRESRSDQLKRHRHGVKPFQCKTCQKRKFSRSDHLKTYHTRHTGKTSE 411
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Search completed: March 1, 2001, 16:16:33
Job time: 413 sec

